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Chondrosarcoma of the Temporomandibular Joint: A Case Report and Review of the Literature

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Chondrosarcomas (CHSs) are defined by the World Health Organization as malignant tumors with a pure hyaline cartilage differentiation and are therefore characterized by the formation of cartilage, not bone, originating from neoplastic cells.[1 and 2]CHSs have been widely reported in the literature and may arise in any region where cartilage is present.[3, 4, 5, 6 and 7]

Depending on the author, 5% to 12% of CHSs are confined to the head and neck region. They represent fewer than 5% of all tumors in this area and are therefore considered very rare.[8, 9 and 10]As reported by Sammartino et al, 49 cases, 14 in India and 35 in Japan, have been described over a period of 50 years.[11]In a study of 56 maxillofacial CHSs over a period of 65 years, Saito et al listed the involved sites and the percentage of presentation as follows: the maxilla (44.6%); the nasal septum, the ethmoid, and sphenoid bone (41.1%); the nose tip (3.6%); and the mandible (10.7%).[12]Within the mandible, the main affected areas are those with an enchondral ossification as opposed to those with a membranous ossification (corpus and ramus).[13 and 14]This intuitively led to the hypothesis that CHSs may arise from mesenchymal cells or from embryonic remnants of the cartilage matrix.[15 and 16]

CHS of the temporomandibular joint (TMJ) is instead extremely rare. In the literature only 17 cases have been reported to date (Medline 1966-2009, English language only; Table 1).[3, 17, 18, 19, 20, 21, 22, 23, 24, 25, 26, 27, 28 and 29]In this report, the authors present 1 case of condylar CHS along with a review of the existing literature.

Table 1.

Reported Cases of TMJ Chondrosarcoma

First Author	Gender	Age	Duration of Symptoms (mo)	MainComplaint	Imaging	Surgery	Irradiation	Follow-Up
Gingrass (1954)[17]	F	46	12	Swelling, pain	Slight TMJ space widening, subcortical sclerosis	X		—
Lanier (1971)[18]	F	48	24	Swelling, pain	Condylar resorption	X		Fewmonths
Richter (1974)[19]	M	75	10	Swelling, pain	TMJ space widening, erosion of the glenoid fossa,	X		12 m

First Author	Gender	Age	Duration of Symptoms (mo)	MainComplaint	Imaging	Surgery	Irradiation	Follow-Up
					increased condylar length			
Tullio (1974)[20]	F	17	8	Swelling	Condylar resorption	X		—
Nortje (1976)[21]	M	40	6	Dullpain, swelling	TMJ space widening, distortion and increased condylar length	X		24 m
Sato (1977)[3]	—	—	36	Pain	—	X	X	—
Sato (1977)[3]	—	—	18	Swelling, pain, trismus	Radiopacity	X		—
Sato (1977)[3]	—	—	4	Pain, trismus	—	X		—
Cadenat (1979)[22]	F	60	—	Swelling, pain	—	X		6 m
Morris (1987)[23]	F	29	24	Swelling, headache	Calcification, mass from condyle to infratemporal fossa	X	X	6 m
Wasenko (1990)[24]	F	49	—	Swelling, pain	Calcification, mass from condyle to infratemporal fossa	X		—
Nitzan (1993)[25]	F	36	72	Swelling, pain	Condylar resorption, radiolucency of TMJ space lesion	X		7 yr
Sesenna (1998)[26]	F	60	12	Swelling	Calcification, mass from condyle to infratemporal fossa	X		5 yr

First Author	Gender	Age	Duration of Symptoms (mo)	Main Complaint	Imaging	Surgery	Irradiation	Follow-Up
Mostafapour (2000)[27]	F	31	96	Swelling	Left pterygoid mass with involvement of TMJ	X		—
Mostafapour (2000)[27]	F	52	18	Swelling	Mass centered on TMJ with involvement of petrous temporal bone and middle fossa	X	X	6 m
Kyoung-in (2008)[28]	F	29	120	Trismus, laterodeviation	Mass centered on TMJ with condylar resorption	X	X	—
Gallego (2009)[29]	M	54	3	Swelling, pain, trismus	Mass from condyle to infratemporal fossa	X		16 m
Garzino-Demo (our case) 2008	F	65	3	Swelling, pain	Mass centered on TMJ with condylar resorption and calcification	X	X	9 yr

REPORT OF A CASE

A 65-year-old female patient affected by a hard and painful mass adjacent to the right condyle was referred to the Division of Oral and Maxillofacial Surgery, University of Turin Hospital. The patient mainly complained of persistent swelling and progressive dull pain for the previous 3 months. Clinical examination showed that mouth opening and lateral and protrusive movements were normal.

Panoramic radiography showed osteosclerotic changes of the right condyle, the shape of which was altered from erosion. Amorphous and slightly radiopaque structures were detectable ahead of the condyle (Figure 1 and Figure 2).

Computed tomographic (CT) scan confirmed and better demonstrated osteosclerosis and erosion of the condyle and clearly depicted a nonenhancing mass with mottled calcification surrounding the condyle itself. Such a mass was grossly modeled on the articular capsule in which it appeared to be confined. The overall radiographic appearance suggested the presence of an intra-articular tumor

with a calcified component due to both bone erosion and new formation of mineralized tissue compatible with CHS (Figure 3 and Figure 4).
An initial ultrasound-guided fine-needle aspiration biopsy (US-FNAB) was inadequate for diagnosis, whereas a second preoperative FNAB led to the diagnosis of a low-grade CHS (Figs 5A-C). Although chances of metastasis were slight, a scintigraphy was performed anyway and showed an uptake in the right TMJ region (Fig 6).
Following a superficial parotidectomy and identification of the main branch of the facial nerve, as well as the zygomatic and buccal branches, a wide radical excision with a zygomatic arch osteotomy and a resection of the upper part of the mandibular ramus was performed. All structures of the TMJ---including the articular capsule and disc---were sacrificed with the exception of the glenoid fossa (Figure 7 and Figure 8). An arthroprosthesis with a titanium condyle head was inserted, and the surgical gap was filled at the glenoid fossa with the posterior half of a temporal myofascial flap (Figure 9 and Figure 10). Postoperative histopathological examination confirmed the diagnosis of a grade I CHS (Fig 1D). On the fifth day, the patient was discharged and later received radiation therapy (74 Gy). Nine years elapsed without any sign of recurrence.

DISCUSSION

The incidence of TMJ neoplasia is very low. In this region, the presentation mode of tumors may mimic those symptoms related to much more frequent pathologies, such as chronic inflammation. In particular, chronic and progressive limitation in mouth opening, swelling, and laterodeviation may be caused by masses such as benign or malignant tumors and synovial chondromatosis.
In a review of TMJ tumor masses, Mostafapour and Futran highlighted an average diagnostic delay ranging from 13 months to 8 years.[27]Moreover, an initial misdiagnosis is frequent. The authors thus suggest further instrumental investigation in the case of progressive difficulty in mouth opening, laterodeviation, and swelling in the preauricular region lasting for more than 1 month and not responsive to therapy.
Mandibular CHS especially when involving the TMJ, is extremely rare. The molar region and, in descending order, the symphysis and the coronoid process may be involved more frequently.[4, 13 and 14]

CHS may be primitive or secondary depending on whether it develops ex novo or from pre-existing lesions (enchondroma, osteochondroma). It may have an endosseous origin---central CHS---or arise from the periosteum-iuxtacortical CHS.[11]Only 17 cases of TMJ CHSs have been reported so far in English (Medline 1966 to January 2009). Review of the literature shows that there is a clear prevalence in females (11 of 17 were female, in 3 cases gender was not stated) and this is in conflict with the overall facial skeletal occurrence where the affected males are 60% and the affected females are 40%.[4 and 30]
The age spectrum ranges from 17 to 75 years (mean, 36.8). The diagnostic delay goes from 3 months to 12 years; this is quite similar to what Mostafapour and Futran reported in their revision of TMJ tumor masses.[27]The delay is basically due to a misidentification of CHS with TMJ chronic inflammatory pathologies.
The most commonly recognized symptom is a preauricular swelling (14 of 17 cases), followed by spontaneous pain as well as pain during mastication (11 of 17), whereas trismus (4 of 17) and laterodeviation at mouth opening (1 of 17) are quite rare. As Mostafapour and Futran pointed out, CHS is generally more painful than enchondroma.[27]
Conventional radiographs may often afford evidence of chondrosarcoma. Pathognomonic signs are the presence of an irregular erosion of the condyle with calcifications localized within the articular space. However, they do not provide adequate information on the extension of the lesion; therefore, other imaging modalities such as CT or magnetic resonance imaging should be performed to confirm the diagnosis and to complete the preoperative workup. With these techniques a

nonenhancing mass with flocculent calcification can be seen at the level of the condyle affected, with or without osseous destruction. The mass may contain calcifications, and this was the case in 5 of 17 patients. It may cause deformities (2 of 17) and condylar erosions (5 of 17). In most cases an increase in the articular space is observed (10 of 17) as well as an involvement of the extra-articular structures (6 of 17) with an erosion of the glenoid fossa and involvement of the infratemporal fossa along with the temporal bone and the middle cranial fossa.

Performing both magnetic resonance imaging and CT are indicated to achieve the best treatment plan and, although metastatic potential is low on average, scintigraphy can be one further examination to carry out.

Histologically, it is possible to observe the proliferation of hyaline cartilage and the presence of a sarcomatousstroma, which contains star-shaped cells, spindle-shaped cells, and round cells.[31]Histopathological CHS classification is based on tumor grading. Evans et al classified CHS in 3 separate grades according to the number of mitoses, cellularity, and tumor size.[32]Grade I shows hypercellularity, chondrocytes with an increased volume, and polymorph hyperchromaticbinucleate forms. Some cases, mostly large lesions, may have a mixoid matrix with foci of calcification. There is a slight to moderate atypia but mitoses are quite rare. Grades II and III show a progressive increase in nuclear polymorphism and the presence of numerous large nuclei.

Cellular differentiation affects metastatic potential. Metastases rates range from 10% in grade II CHS to 71% in grade III CHS. No metastases are reported in grade I CHS.[26]These data are, however, related to bones and joints of other parts of the body.

Local recurrences are instead independent of tumor grading and seem to be related to inadequate previous surgical therapy.[26]

Data concerning TMJ CHSs are not complete because they are not always fully reported and are sometimes lacking information about tumor grading. Also, many authors report intermediate grades between I and III. However, when grading is stated, grade I CHSs appear to be prevalent.

Regarding the diagnostical phase, biopsy of CHS is not advised by some authors because of the risk of anaplastic transformation and diffusion during manipulation of the mass.[33 and 34]Moreover, as Morris et al pointed out, FNAB does not always provide a reliable diagnosis.[23]Here FNAB was initially inadequate for diagnosis but a second cytology actually led to the correct diagnosis, which was then confirmed by conventional histopathological examination. Incisional biopsy is, however, the best option for distinguishing among osteogenic sarcoma, pleomorphic adenoma, and chondroma.

Because of the rarity that maxillomandibular CHS represents, there are no specific treatment protocols outlined. This explains that treatment modalities are the ones that have been previously used for CHS and sarcomas of other regions.

Surgical therapy represents the gold standard for primary treatment of this neoplasm.[7, 35, 36 and 37]Resection must be as wide as possible, and the presence of large healthy tissue margins (>2 to 3 cm) seems to positively affect prognosis and chances of recurrence.[11 and 36]

Other treatment modalities include intralesional resection and curettage along with radiation therapy. These modalities are not curative and have been described for large lesions that cannot possibly be treated with surgery only.[35 and 38]

Cryotherapy has been described for grade I lesions.[39, 40 and 41]This treatment may provide advantages from a functional point of view but not as many from the oncologic one.

In all 17 cases reported in the literature and in the present case, CHS was treated by surgery. In a few cases, surgery implied emimandibulectomy,[20 and 21] but more frequently a simple condylectomy was performed.[23, 25 and 28]In more extended cases, such as the ones described by Mostafapour and Futran, surgery consisted of frontotemporal craniotomy and cranial base resection followed by reconstruction with rectus abdominis free flaps.[27]

Although resection seems the surgical technique of choice for maxillomandibular CHSs, a high percentage of recurrence has been described with surgery alone.[11]Therefore, a combination of surgery and radiation therapy and/or chemotherapy has been proposed. However, results on

recurrences and survival still appear controversial. Radiation therapy alone is not effective, but its use along with surgery seems to increase survival rate and local control of the disease. More specifically, adjuvant radiation therapy in TMJ CHS has led to a long survival period in 3 of 17 reported cases, and the same holds true for the case presented here as well.[14 and 32]

The choice of not using a complete fossa and condylar head prosthesis was determined by the fact that our patient had to undergo radiation therapy. Our experience with complications due to the presence of metal prosthetic devices in radiotherapeutic patients made us opt for a condylar head prosthesis.

Once again it is important to highlight the fact that survival rates are often not stated and that, although most surgeons actually recommend this sort of therapy, some of their patients may not undergo full treatment for their own personal reasons.

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